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# Prosopamnesia: a case report of amnesia for faces

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#### ABSTRACT

Prosopamnesia is a face-selective memory disorder in which face learning is impaired, while faceperception disorder (prosopagnosia) and memory disorders for stimuli other than faces are not present. To date, only two cases of prosopamnesia have been reported in adults – one congenital and one secondary to brain damage. This article reports a case of a 68-year-old woman complaining difficulties recognizing persons she had got to know recently. Neuropsychological examination revealed facespecific anterograde amnesia in the absence of prosopagnosia and other memory impairments. Brain MRI did not present any focal abnormality; PET-scan revealed hypoactivation mostly in the frontotemporal area bilaterally. This patient represents the first case of late-onset primary prosopamnesia. ARTICLE HISTORY Received 19 September 2021

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**KEYWORDS** 

Prosopamnesia; prosopagnosia; amnesia; memory; face learning; face processing; face recognition

# Introduction

#### Material-specific memory impairment

Several cases of material-specific amnesia with damage in discrete brain regions have been reported. For example, visual memory impairment that involves the right hemisphere and unilateral lesions of the medial-temporal lobe (Milner, 1971; Pigott & Milner, 1993) or verbal memory impairment in cases of left-sided lesions (Frisk & Milner, 1990). An extended level of selectivity can be identified in cases of class-specific amnesia. Numerous studies regarding visual memory have reported the following: selective topographical memory impairment in which no other aspects of visual memory were affected (Incisa Della Rocchetta et al., 1996; Maguire et al., 1996), a selective visual memory deficit involving face and animals with no impairment of topographical memory (Cipolotti et al., 1999), verbal and topographical memory impairment with a spared memory for faces (Cipolotti et al., 2006), a topographical memory deficit with no impairment of verbal or face-specific memory (Bird et al., 2007), and a selective sparing of topographical memory in the context of a severe memory impairment for any other stimuli (Maguire & Cipolotti, 1998). Furthermore, a case of global amnesia in which only face-specific memory was spared has been reported (Carlesimo et al., 2001).

#### Face memory impairment: prosopamnesia

Prosopagnosia and prosopamnesia are two neurocognitive disorders involving face processing and refer to impairment of facespecific perception and face-specific memory, respectively. Prosopagnosia is caused by cortical lesions affecting the occipitaltemporal areas, frequently due to ischemic insult of the right posterior cerebral artery territory. In contrast, prosopamnesia remains to be fully defined as a syndrome. It refers to a selective memory disorder for faces, characterized by an impairment in face learning in the absence of prosopagnosia and memory impairment for stimuli other than faces. Prosopamnesia has been defined by Tippett et al. (2000) as "the most selective material-specific anterograde amnesia yet described" and highlights a disconnection between the perceptual domain (i.e., visual coding of the "face" stimulus) and memory (i.e., storage of the face over time).

# **Clinical features**

According to Tippett et al. (2000) the diagnosis of prosopamnesia can be formulated when the following conditions are present:

- (1) Adequate performance on face perception tasks;
- (2) Poor performance on learning tasks involving faces;

(3) Adequate performance on learning tasks involving visual stimuli other than faces;

(4) Intact recognition of faces of people encountered prior to the brain injury.

# Cases reported in the literature

To date, two confirmed cases of prosopamnesia have been reported: one with symptoms that started after a right temporal lobectomy in a patient with refractory seizures following a head injury (Tippett et al., 2000), and the second case presented a congenital form of prosopamnesia (Williams et al., 2007). A third case has been reported in the literature describing a patient that was defined as "prosopamnesic" after a head trauma (Lopera & Ardila, 1992). However, this patient did not fulfill the above criteria for prosopamnesia as he also reported prosopagnosia and visual memory impairment.

# Methods

This study reported a case of late-onset prosopamnesia not due to focal brain injury. The patient is a Caucasian Italian woman, who was 68 years old at the time of testing, with 8 years of education (graduated from middle school). She presented to

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the Neurology and Neuropsychology outpatient service complaining of a 4 – 5-year history of difficulties with recognition of persons that she had got to know recently. She said that this problem becomes more evident when she reencounters someone she has already known in a different context or situation from the first meeting. The patient lives in a 25,000 inhabitants' town. She reported that while she is walking, often someone she does not apparently know greets her or sometimes stops and talks to her. In this latter case she feels uncomfortable and tries, through the conversation, to intercept any useful information to understand who they are. She did not have any complaints regarding difficulties in recognizing people from her family, close-relatives, and long-term friends. Although this symptom remained stable through time and did not limit her daily activities, it impacted her social life.

The patient was neurologically examined in June 2017, and she received the first neuropsychological assessment in May 2017, with a follow-up in November 2017. The following areas were investigated using specific neuropsychological tests: global cognition (Mini Mental State Examination (Magni et al., 1996)), visual attention (Multiple Features Target Cancellation test (Marra et al., 2013)), short-term and working memory (Digit Span and Corsi Block-Tapping Task (Monaco et al., 2013)), episodic long-term memory (Rey Auditory Verbal Learning Test (Carlesimo et al., 1996), Rey-Osterrieth Complex Figure (Caffarra et al., 2002), Corsi Block-Tapping Supra-Span Task (Spinnler & Tognoni, 1987)), executive functions (Frontal Assessment Battery (Appollonio et al., 2005), Raven's Colored Progressive Matrices (Basso et al., 1987), Phonemic Fluency and Semantic Fluency (Costa et al., 2014), Clock Drawing Test (Mondini et al., 2011)), and lexicon retrieval (15-Picture Naming Test (Della Sala et al., 1996)).

To assess face processing abilities, apperceptive feature processing (Physiognomic Decision Task (Della Sala et al., 1995) and associative feature processing (12-Famous-Face Naming test (Bizzozero et al., 2007), 90-Recent-Famous-Face Recognition task) were tested. Face learning was examined with the Recognition Memory Test (RMT, Smirni et al., 2010). The Physiognomic Decision Task (Della Sala et al., 1995) is a test in which 20 stylized visual patterns are presented, 10 resembling a face and 10 resembling a non-face; for each of these patterns, subjects have to report whether they see a face (Figure 1).

The 12-Famous-Face Naming test (Bizzozero et al., 2007) is a standardized confrontation naming task involving 12 internationally famous characters belonging to three different historical periods (pre World War II, 1950–1970's, and 1980– 1990's), from three categories (show business, culture, and politics) (Figure 2).

The 90-Recent-Famous-Face Recognition task is an ad hoc clinical task that we created to assess the recognition of more recent characters than the previous test. It comprises 90 faces of Italian and non-Italian individuals who were popular in a particular period (1980's, 1990's, 2000's, and post 2011), and belonged to different categories (show business, sports, culture, and politics). The test comprises two tasks: 1) identification, where the patient has to provide a semantic description of the character (what was the popular individual's job, the reason for which they were famous), and 2) naming. If the patient was unable to learn new faces since the onset of her symptoms (that she dates since 2011–2012), then she should have difficulties in recognizing faces from recent years (the ones whose gained popularity after 2011) and no difficulties in recognition faces from the previous decades (1980's, 1990's, and 2000's).

The Recognition Memory Test (RMT, Smirni et al., 2010) is an Italian test used to assess material-specific memory deficits. It comprises three subtests: recognition memory for words (4–6 letters middle frequency Italian words, RMT – words), recognition memory for unfamiliar faces (RMT – Faces), and recognition memory for unfamiliar buildings (RMT – Buildings). Each subtest is composed of 30 stimuli printed singularly on a A4 page that are presented to the subjects at 3-second intervals. For each stimulus, the subject must state whether they like it. The learning phase is immediately followed by the recognition phase that utilizes forced-choice recognition methodology with three similar alternatives (Figures 3-4).





Figure 1 Examples of stimuli used in the Physiognomic Decision Task (Della Sala et al., 1995).



Figure 2 Examples of stimuli used in the 12-Famous-Face Naming test (from the left: the American president John Fitzgerald Kennedy, the Italian composer Giuseppe Verdi, the Italian actor Roberto Benigni and the American actor Oliver Hardy; Bizzozero et al., 2007).



Figure 3 Examples of stimuli used in the RMT - Faces. A: learning phase; B: recognition phase (Smirni et al., 2010).

Neuropsychiatric symptoms were assessed using the Neuropsychiatric Inventory Questionnaire (NPI-Q). Brain magnetic resonance imaging (MRI) and 18 F-fluoro-deoxy-glucose positron emission tomography (FDG-PET) scan were performed as well.

# Results

#### **Clinical examination**

The neurological exam was unremarkable, except for an essential tremor of the head. The neuropsychological assessment revealed an intact global cognitive profile with the highest score on the global functioning test (MMSE score: 30/30; Table 1).

In function-specific tests her performance was normal, with scores above the twentieth percentile of the normal population. The only exception was the RMT for Faces (Smirni et al., 2010), which assesses learning skills for unfamiliar faces. The patient's performance was pathological, with a score below the fifth percentile. On the 90-Recent-Famous-Face Recognition task, the ad-hoc task assessing the recognition of famous faces, her performance was good for characters belonging to the first three decades (1980's, 1990's, and 2000's) and lower for characters of the most recent years (post 2011) (being an ad hoc clinical task, normative data are not present). NPI-Q did not indicate any behavioral disorder or emotional imbalance.



Figure 4 Examples of stimuli used in the RMT - Buildings. A: learning phase; B: recognition phase (Smirni et al., 2010).

| Table 1. First neuropsychological assess | ment results                      |          |              |  |
|--|-----------------------------------|----------|--------------|--|
|  | Score                             | Perc     | entile       |  |
| MMSE                                     | 30                                | >        | •5           |  |
| MFTC                                     | 11                                | >        | •5           |  |
| Digit Span Forward                       | 6                                 | >        | 50           |  |
| Digit Span Backward                      | 4                                 | >        | 50           |  |
| Corsi Block-Tapping Task                 | 4                                 | 21 -     | - 35         |  |
| Corsi Block-Tapping Supra-Span Task      | 17.17                             | >50      |              |  |
| RAVLT – Immediate Recall                 | 45                                | >50      |              |  |
| RAVLT – Delayed Recall                   | 10                                | >50      |              |  |
| ROCF – Copy                              | 33                                | >        | >50          |  |
| ROCF – Delayed Recall                    | 20                                | >        | 50           |  |
| FAB                                      | 16                                | >        | 50           |  |
| Raven's CPM                              | 20                                | 21 -     | - 35         |  |
| Phonemic Fluency                         | 45                                | >        | 50           |  |
| Semantic Fluency                         | 48                                | >50      |              |  |
| CDT                                      | 9                                 | >        | •5           |  |
| 15-Picture Naming Test                   | 13                                | >5       |              |  |
| Physiognomic Decision Task               | 17                                | >5       |              |  |
| 12-Famous-Face Naming                    | 73.5                              | >50      |              |  |
| RMT – Words                              | 28                                | >50      |              |  |
| RMT – Faces                              | 17                                | <        | :5           |  |
| RMT – Buildings                          | 22                                | 25 -     | - 50         |  |
| 90-Recent-Famous-Face Recognition        |                                   |          |              |  |
| - 2011-2017                              | Identificatio                     | on 15/23 | Naming 14/23 |  |
|  | (67                               | '%)      | (61%)        |  |
| - 2001-2010                              | Identification 20/23 Naming 18/23 |          |              |  |
|  | (86                               | 5%)      | (76%)        |  |
| - 1991-2000                              | Identification 21/22 Naming 19/22 |          |              |  |
|  | (96%) (85%)                       |          | (85%)        |  |
| - 1981-1990                              | Identification 20/22 Naming 17/2  |          | Naming 17/22 |  |
|  | (92%)                             |          | (75%)        |  |

MMSE, Mini Mental State Examination; MFTC, Multiple Features Target Cancellation; RAVLT, Rey Auditory Verbal Learning Test; ROCF, Rey-Osterrieth Complex Figure; FAB, Frontal Assessment Battery; CPM, Colored Progressive Matrices; CDT, Clock Drawing Test; RMT, Recognition Memory Test.

# Neuroimaging results

Brain MRI did not reveal any focal areas of altered signal intensity in the cerebral hemispheres, brainstem or cerebellum; ventricular system appeared normal; cortical subarachnoid space at vertex was enlarged; and there was no evidence of intracranial space occupying lesion. Brain FDG-PET scan presented evidence of reduced tracer uptake in the fronto-temporal areas bilaterally as well as in the left parietal lobe, although to a lower extent (Figure 5). Possible signs of chronic vascular lesions were observed; however, these alterations were not considered specific of a particular pathological condition.

#### **Clinical follow-up**

Six months after the initial examination, the neuropsychological assessment showed similar test results with an unchanged cognitive profile (Table 2). Once again, the RMT for Faces (Smirni et al., 2010) revealed a pathological performance, where the patient's score decreased from 17/30 in the first assessment to 14/30. The 90-Recent-Famous-Face-Recognition task performance continued being lower for individuals becoming famous post 2011.

#### Discussion

This study described a patient with a selective memory impairment for faces. She had an intact global cognitive profile, with normal performance in all the neuropsychological tests except an isolated pathological performance on the RMT for Faces (Smirni et al., 2010). In addition, she exhibited difficulties in recognizing celebrities who became famous after 2011, compared to the ones who rose to fame before 2011. This evidence has been confirmed at the 6 months follow-up assessment.

This clinical picture fulfills the criteria for prosopamnesia defined by Tippett et al. (2000) as the patient exhibited:

• Spared visual processing abilities, including face perception. The optimal performances of the visual searching, complex figure copy, picture naming, and physiognomic decisionmaking tasks allowed us to conclude that the patient was capable of proper elaboration of all the visual stimuli presented

(i.e., faces, objects, and shapes).

• Impaired face learning.

The patient's poor performance on the RMT for Faces on both first and follow-up assessment indicates an inadequate learning of the stimulus face. Her normal performances on the RMT for Words and on the RMT for Buildings indicates that maintaining the same testing procedure but with other kind of stimuli besides faces learning is adequate.

• Spared learning of other types of stimuli besides faces.

In addition to RMT for Words and for Building the patient's performance was fully normal in memory tests for visuo-spatial sequences, complex figure, and list of words. Thus, her episodic memory proved to be completely normal for all the non-facial tested stimuli.

Spared recognition of faces known prior to the disease.

The patient did not report problems in recognizing familiar faces, such as members of her family, close relatives, and long-term friends. These "stimuli" were acquired before the onset of her symptoms. Her normal performance on the 12-Famous-Face Naming suggests an overall preserved face recognition of the celebrities of the last century. To explore the recognition of recent celebrities, the ad-hoc 90-Recent-Famous-Face Recognition task has been created. Given that the task does not have normative data, it is inconclusive whether the lower performance with faces post 2011 can be considered pathological. Clinically, these findings indicate that the patient's recognition of faces known to her prior to the disease was adequate, whereas she had some problem in recognition of faces to which she was exposed after the onset of her symptoms.

The features presented above suggest that the patient's dysfunction was of an anterograde amnesia type, specific for faces (prosopamnesia), with spared face perceptual processing (she did not exhibit apperceptive prosopagnosia) and spared pre-onset face recognition (she did not exhibit associative prosopagnosia).

| Table 2. Follow-up n | neuropsychologica | l assessment results |
|----------------------|-------------------|----------------------|
|----------------------|-------------------|----------------------|

| 1 1, 3                              |   |       |                                 |  |
|-------------------------------------|---|-------|---------------------------------|--|
|                                     | Score                                   | Perce | entile                          |  |
| MMSE                                | 29                                      | >     | -5                              |  |
| MFTC                                | 13                                      | >     | >5                              |  |
| Digit Span Forward                  | 6                                       | >     | >50                             |  |
| Digit Span Backward                 | 4                                       | >     | >50                             |  |
| Corsi Block-Tapping Task            | 6                                       | >     | >50                             |  |
| Corsi Block-Tapping Supra-Span Task | 14.32                                   | 36-   | 36–50                           |  |
| RAVLT – Immediate Recall            | 48                                      | >     | >50                             |  |
| RAVLT – Delayed Recall              | 11                                      | >     | >50                             |  |
| ROCF – Copy                         | 33                                      | >     | >50                             |  |
| ROCF – Delayed Recall               | 21                                      | >     | >50                             |  |
| FAB                                 | 18                                      | >     | >50                             |  |
| Raven's CPM                         | 22                                      | 21 -  | 21 – 35                         |  |
| Phonemic Fluency                    | 53                                      | >     | >50                             |  |
| Semantic Fluency                    | 51                                      | >50   |                                 |  |
| CDT                                 | 8.5                                     | >5    |                                 |  |
| 15-Picture Naming Test              | 14                                      | >5    |                                 |  |
| Physiognomic Decision Task          | 18                                      | >5    |                                 |  |
| 12-Famous-Face Naming               | 73.5                                    | >     | >50                             |  |
| RMT – Words                         | 29                                      | >     | >50                             |  |
| RMT – Faces                         | 14                                      | <5    |                                 |  |
| RMT – Buildings                     | 25                                      | >50   |                                 |  |
| 90-Recent-Famous-Face Recognition   |   |       |                                 |  |
| - 2011-2017                         | Identification 15/23 Naming 13/23       |       |                                 |  |
| - 2001-2010                         | Identification 21/23 Na                 |       | Naming 18/23                    |  |
| - 1991-2000                         | Identification 21/22                    |       | (70%)<br>Naming 18/22           |  |
| - 1981-1990                         | Identification 20/22 Naming<br>(92%) (7 |       | (8170)<br>Naming 17/22<br>(75%) |  |
|                                     |   |       |                                 |  |

MMSE, Mini Mental State Examination; MFTC, Multiple Features Target Cancellation; RAVLT, Rey Auditory Verbal Learning Test; ROCF, Rey-Osterrieth Complex Figure; FAB, Frontal Assessment Battery; CPM, Colored Progressive Matrices; CDT, Clock Drawing Test; RMT, Recognition Memory Test.

This dysfunction has been highlighted through memory tests of both unfamiliar faces (RMT for faces) and familiar/famous faces (part 2011–2017 of the 90-Famous-Face-Recognition task).

Carlesimo et al. (2001) reported a patient that was presented with global amnesia with selective sparing of face learning after acute carbon monoxide poisoning. MRI of the patient's brain revealed severe bilateral hippocampal atrophy. Our patient represents a double dissociation compared to this case. Based on these findings, it may be speculated that face learning is independent of the hippocampus. This hypothesis is also supported by numerous studies on brain damaged patients, healthy subjects, and animals, which considered non-hippocampal regions to be neural areas involved in face perception and learning (Bird, 2017; Gobbini & Haxby, 2007; Kanwisher, 2017).

Several studies on the encoding and recognition of previously unfamiliar human faces (i.e., without biographic or semantic components) reported that fusiform gyrus activation was bilateral during face encoding and lateralized to the right



Figure 5 A brain FDG-PET scan of the patient revealed hypometabolism in the fronto-temporal region bilaterally and slightly in the left parietal lobe.

during recognition (Maguire et al., 2001). The PET scan of the patient revealed bilateral temporal hypometabolism along with frontal involvement. This pattern is non-specific for any particular pathology. However, it does not reject a bilateral involvement of temporal areas that, according to previous studies, could indicate that her face memory dysfunction was related to an impairment in the stage of encoding, and not of retrieval. This interpretation could also be supported by the current behavioral finding. In the face recognition memory task (RMT for Faces (Smirni et al., 2010)), the patient did not benefit from the alternatives displayed, suggesting an inadequate initial acquisition/encoding of information.

To date, only two other cases of prosopamnesia have been reported in adults. One case was secondary to a brain lesion (Tippett et al., 2000) and the other case was a congenital disorder (Williams et al., 2007). In both cases, the topographical memory (memory involved in spatial navigation) was not assessed; thus, it is unclear whether their visual memory impairment was limited only to faces. In the current case, performance on learning tests of buildings, complex figure, and visuospatial sequences was completely intact. Based on these results, the patient presented all the clinical features of prosopamnesia defined by Tippett et al. (2000), where visual memory for stimuli other than faces should be completely spared.

The patient of this study represents the first reported case of late-onset primary prosopamnesia as her symptoms were not related to focal brain insults (e.i., trauma, inflammatory, vascular, or infectious disease). Further follow-ups will enable us to verify whether this will remain an isolated deficit or will progress into a broader progressive disease.

# **Authors' contributions**

MPG conceived and planned the study. MB performed the neurological examination. SM performed the neuropsychological assessment, data collection, and interpretation. IMS supervised the entire study. SM produced the first draft of the manuscript in consultation with MPG. All authors read, provided input, and approved the final manuscript.

# **Ethics approval**

Approval by the Ethics Committee was not required as this was a singlecase report based on data from standard clinical practice.

#### Data availability statement

The authors confirm that the data supporting the findings of this study are available within the article.

#### **Disclosure statement**

No potential conflict of interest was reported by the author(s).

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